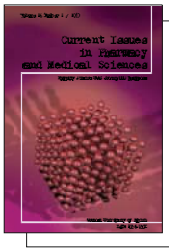

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Caroli disease in magnetic resonance imagining

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ABSTRACT

Caroli disease is a rare liver disorder, characterized by saccular or fusiform multifocal, segmental dilatation of the intrahepatic bile ducts. A new case of the disease diagnosed in a 38-years old male using magnetic resonance with virtual cholangiopancreatography is reported.

Keywords: Caroli disease, bile ducts, hepatic cirrhosis, magnetic resonance

INTRODUCTION

Cyst or cystic-like lesions are common findings during ultrasound and various radiological examinations of the liver. Such abnormalities include simple cyst, pyogenic abscess, echinococcal cyst, amebic abscess, metastasis, biliary cystadenoma and cystadenocarcinoma, hepatocellular carcinoma and cholangiocarcinoma. They could be also visible in adult polycystic kidney disease, and extrapancreatic pseudocyst [7, 13]. The last group contains also a rare Caroli disease which is characterized by a congenital cystic dilatation of the intrahepatic bile ducts. Presently, it is divided into a simple and complex type. The first one, also known as isolated form, is secondary to the local bile duct(s) ectasia and dilatation of peripheral bile system in an affected part of the liver. The second type – so-called Caroli syndrome – coexists with a portal hypertension and congenital hepatic fibrosis [11, 9].

The current report presents a new case of Caroli disease based on magnetic resonance examination.

CASE DESCRIPTION

A 38-year-old male with abdominal pain and ascites, preliminary diagnosed as hepatic cirrhosis on the basis of utrasonography and computer tomography was submitted to Magnetic Resonance Unit of St. John's Cancer Center (Lublin, Poland).

Magnetic resonance (MR) examination was performed on 1.5T MR scanner (Achieva; Philips Medical Systems; Veenpluis, The Netherlands) using short protocol dedicated for the virtual cholangiopancreatography (MRCP) which included routine TFE-T1-weighted axial, TFE-T1-weighted coronal, TSE-T2-weighted axial and STIR-T2-weighted axial series. The section thickness and intersection gap was constant and kept at 5 and 1 mm, respectively. The radial virtual MRCP (section thickness 1.6 mm, intersection gap 0 mm) was performed. The paramagnetic contrast administration was not applied. Data was stored on the picture archiving and communication system (PACS).

The examination revealed hepatomegaly (242x173 x186 mm; coronal x sagittal x vertical) with signs of the macronodular cirrosis. The organ parenchyma presented irregular shape and intensity in all applied series (fig. 1). Numbers of single regenerative nodules with diameter up to 37 mm were found. Saccular and fusiform dilatations (up to 8 mm) of segmental bile ducts were observed in right and left lobe (fig. 2). Local narrowing of the segmental, right and left hepatic ducts as well as common hepatic and common bile ducts were also presented on MRCP reconstruction.

Additionally, fluid in peritoneal cavity and slight dilatation of the hepatic portal vein (16 mm) were observed. However, morphology of other abdominal organs was unaffected.

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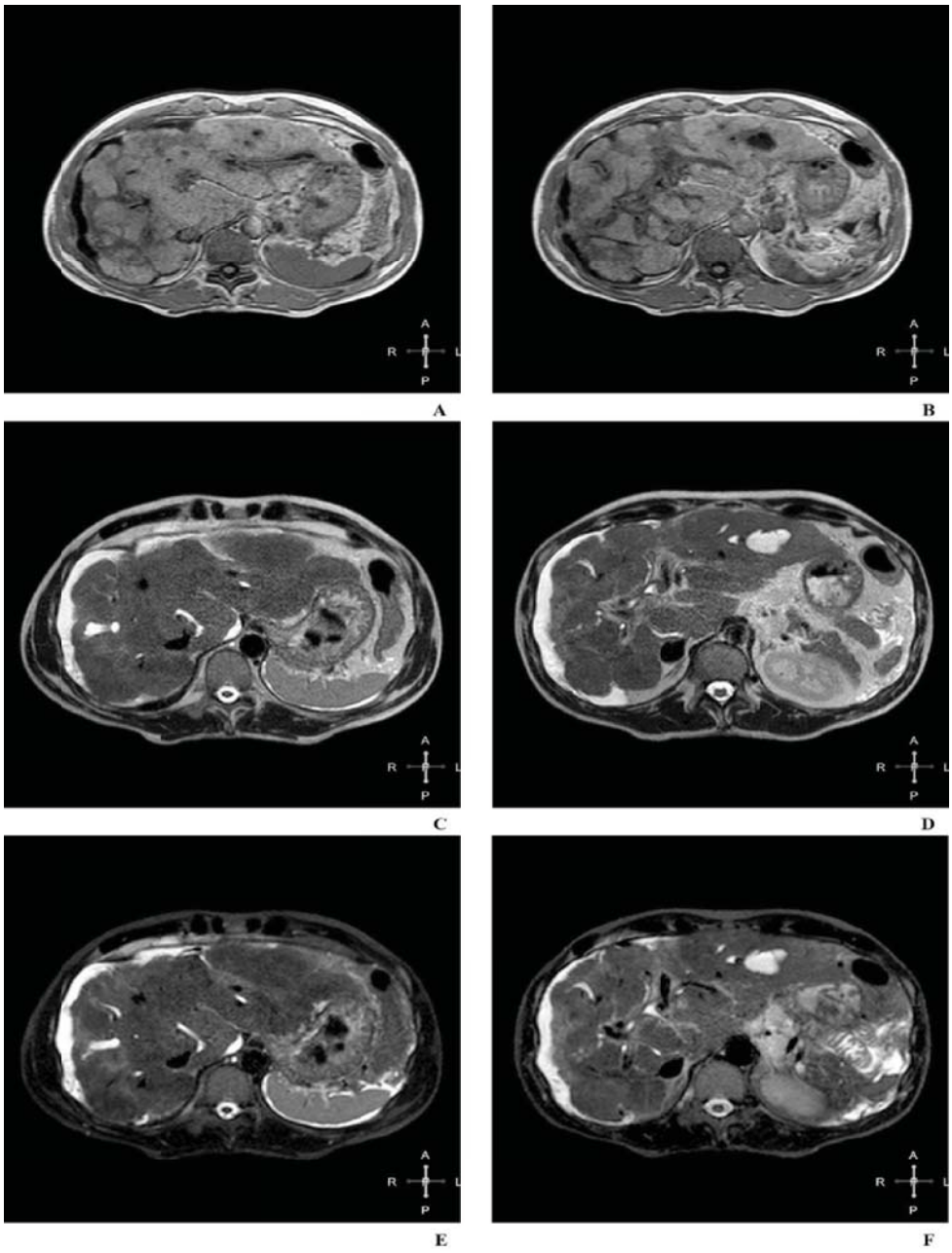

Fig. 1. Dilatation of segmental bile ducts in hepatic right (A, C, E) and left lobe (B, D, F) in patient with liver cirrhosis and ascites. A, B – axial T1-weighted section; C, D – axial T2-weighted axial section; E, F – axial STIR section

Because of relatively good general condition, a symptomatic treatment and radiological follow-up was suggested.

DISCUSSION

The current case describes typical morphology of Caroli diseases [3]; however with a wide dilatation of the segmental bile ducts. Such manifestation is probably age-dependent and fully corresponds to a classic description done by Jacques Caroli (1902-1979) **[1].** Similar morphology should be observed in ultrasound examination and computer tomography. However MR, especially with a cholangiopancreatograpy (MRCP), gives the best view of the entire biliary system. Due to high contrast between fluid (high signal in T2- weighted sequence) and calcified elements (low or lack signal in both T1- and T2-weighted sequence) the method allows precise localization of gallstone, that may complicat ducts obstruction [6].

For unknown reason the ductal dilatation are usually monolobar but rare case of bilobal form – like in the presented case – was also described [3]. Moreover, all cysts that are visible in Caroli disease are classified as type V according to Tadani et al. [1977], which are characterized as saccular or fusiform multifocal, segmental dilatation of the intrahepatic bile ducts. In comparison, type I (true choledochal cyst; 80-90% of cyst-like lesions of the liver) results from reflux of pancreatic secretions into the bile duct via anomaly of pancreaticobiliary junction. Such lesions are present as a fusiform dilation of the extrahepatic duct and are divided into type Ia (dilatation of entire extrahepatic bile duct), Ib (dilatation of focal/segmental extrahepatic bile duct) and Ic (dilatation of the common bile duct portion of extrahepatic bile duct). Type II (bile duct diverticulum; 3%) are saccular outpouchings arising from the supra-duodenal extrahepatic bile duct or the intra-hepatic bile ducts. Type III (choledochoceles, 5%) is characterized by protrusion of a focally dilated, intramural segment of the distal common bile duct into the duodenum. Type IV (multiple communicating intrahepatic and extrahepatic duct cysts; 10%) is divided into type IVa (fusiform dilation of the entire extrahepatic bile duct with extension of dilation of the intrahepatic bile ducts) and IVb (multiple cystic dilations involving only the extrahepatic bile duct).

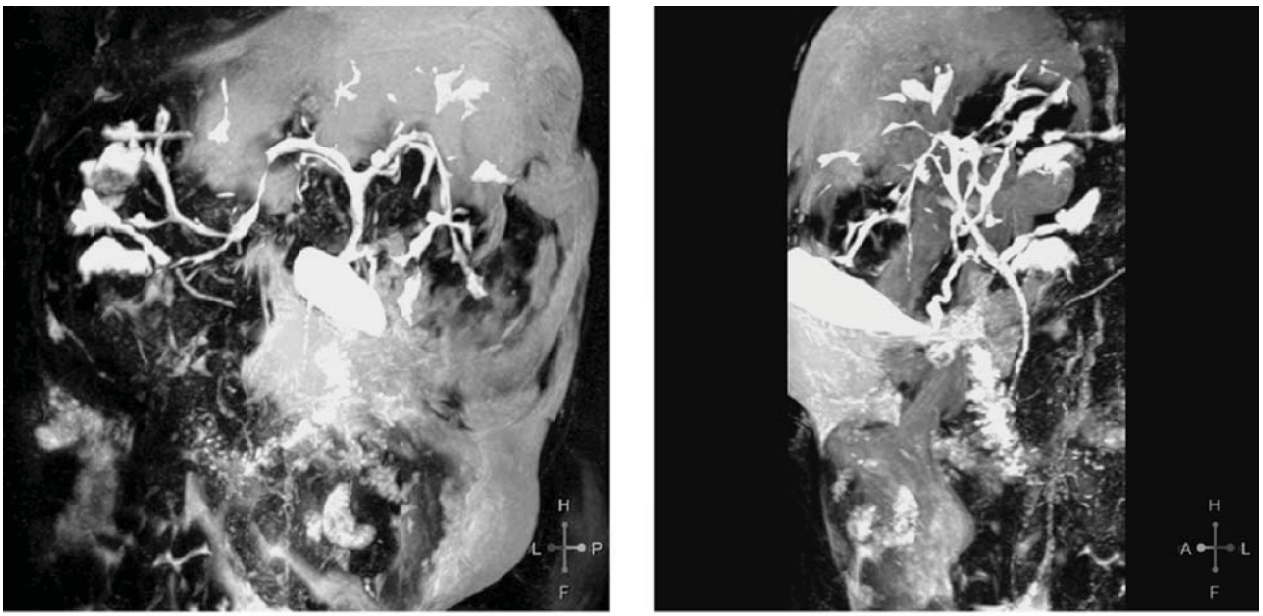

Fig. 2. Dilatation of segmental bile ducts and local obstruction of various pats of intrahepatic bile system in virtual magnetic resonance cholangiopancreatograpy (MRCP). The background shadow is secondary to ascites. A – antero-posterior view; B – right lateral view

Caroli disease is relatively rare since it affects 1 in 1 000 000 people with female predisposition. Type I (isolated form) was found to be autosomal dominant while type II (complex form) is autosomal recessive one. Since autosomal recessive polycystic kidney disease (ARPKD) highly coexists with Caroli disease, mutation of the PKHD1 gene (6p12.2) was also pointed as a target gene for the currently discussed liver pathology [10, 14, 2]. Mutation in WDR19/NPHP13 and GLIS2/NPHP7 genes was also described as a possible case [4]. An apoptosis of biliary epithelial cells related to ductal plate malformation, as well as reduction of laminin and type IV collagen synthesis in the basement membrane of intrahepatic bile ducts were also pointed as an additional factor for the dilatation of bile ducts [8]. Furthermore, virus-induced T-cell mediated autoimmune-mediated cholangiopathy, virus-induced apoptosis of biliary epithelial cells by a TNF-related apoptosis-inducing ligand followed by the progressive obliteration of bile ducts has been currently reported for congenital hepatic fibrosis and biliary atresia that may let to Corali's disease [8].

In spite of etiology and clinical symptoms, it should be stressed that clinical course of Caroli disease may be complicated by portal hypertension, liver cirrhosis, splenomegaly, and cysts of other organs, such as the spleen, kidneys, and pancreas as well as recurrent episodes of cholangitis, frequently with intrahepatic stones as well as infections with gram-negative bacteria that may lead to hepatic abscess formation. However, the disease is also complicated by malignancies, e.g., hepatocellular carcinoma, cholangiocarcinoma [11, 5].

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